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ORBITAL LYMPHOMAS - A CASE SERIES

Hematology

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ABSTRACT

Lymphoma is the most frequent malignancy of the ocular adnexa which may arise in the conjunctiva, eyelids, and orbit including the lacrimal gland. 4 such cases treated in State Cancer Institute, Guwahati is discussed in case series.

KEYWORDS

INTRODUCTION

The orbit of the eye is shaped like a quadrilateral pyramid enclosing the eyeball and structures of the ocular adnexa. Ocular adnexal lymphoma can arise in the conjunctiva, eyelids, and orbit including the lacrimal gland. Lymphoma is the most frequent malignancy of the ocular adnexa ("Orbital Lymphoma," 2019). Lymphomas of the ocular adnexa are a heterogeneous group of malignancies, comprising approximately 1% to 2% of non-Hodgkin lymphomas (NHLs) and 8% of extranodal lymphomas (Møller et al., 2008). In this case series we present 4 patients with newly diagnosed orbital lymphoma that were diagnosed and initiated on treatment at the State Cancer Institute, Guwahati from July 2024 to January 2025.

Case Presentations

Patient 1

62 year old female with no known comorbidities, presented with complaints of left sided frontal headache since 2 months, proptosis of left eye and diminished vision left eye of 3 weeks duration.There was history of unintentional loss of weight (not documented), generalised weakness also. Rest system review was negative. Her initial evaluation was at Regional institute of ophthalmology, Guwahati. CT orbit was done which showed an intraconal space occupying lesion, left orbit. FNAC was suspicious of malignant cells. Biopsy done from the lesion was suggestive of high grade non-Hodgkin lymphoma. She was referred to SCI for further management. On examination her ECOG was 2, positive examination findings included - proptosis of the left eye with exposure keratitis, generalised lymphadenopathy, including bilateral cervical, axillary lymphadenopathy and inguinal nodes. There were also multiple subcutaneous nodules over the anterior and posterior aspect of the chest wall. FDG PET CT whole body showed ill-defined soft tissue in the intraconal compartment of the left orbit compressing the globe causing proptosis measuring 5.5 X 3.7 X 4 cm (SUV max 13.5). There was disseminated disease with nodes both above and below the diaphragm and disseminated soft tissue nodules with high FDG uptake. A repeat biopsy was needed at SCI as the tissue block had been exhausted and immunohistochemistry examination was pending. Repeat biopsy from the chest nodule showed DLBCL, ABC type. She was started on R-CHOP protocol. Her treatment was complicated by chemotherapy extravasation which was grade 3 severity. She was treated with regular wound dressing and topical antibiotics. As there was considerable difficulty in gaining iv access, central line insertion was done from cycle 3 onwards. The interim FDG PET CT that was done post 3 cycles of chemotherapy showed complete metabolic response Post cycle 4 chemotherapy she developed septic shock with the ulceration site as a possible focus of infection.. Post 5 cycles of R CHOP she developed fever with chills. Culture showed pseudomnas aeroginosa which was pan resistent. The central line was removed and she was started on polymixin and meropenem. She further developed drowsiness, seizure episode and aleterd sensorium. Imaging showed mass lesion with perilesional edema. Patient and relatives were not willing for further evaluation or treatement and she was discharged on request on supportive care medications.

Patient 2

45 year old male with no known comorbidities, presented with complaints of swelling right side of the face for 2 months duration, change of voice since 1 month, blocked right nostril with mild recurrent epistaxis for 1 month. No history of fever, chills, drenching sweats, loss of appetite or weight loss. The facial swelling had increased over 2 months causing displacement of the nose and right eye. Epiphora was present. There was also lesion over the palate protruding into the oral cavity.His initial evaluation was at Guahati Medical college under ENT department. Imaging was done by non-contrast CT of the nose and paranasal sinuses which showed, soft tissue lesion involving the right maxillary sinus, right maxillary sinus, right side of ethmoid air cells, right side of sphenoid sinus and extending to right nasal cavity and extraconal compartment of right orbit and resultant proptosis of right eyeball. There was bony erosion seen in the involving bones. Biopsy from the right nasal mass was suggestive of non-hodgkin lymphoma. He was referred to SCI for further management. Slide review and immunohistochemistry of biopsy showed DLBCL, GCB type. FDG PET CT whole body was done for staging and showed FDG avid large heterogeneously enhancing mass involving the right sided nasal cavity, right frontal, bilateral ethmoid, maxillary sinuses, extending into the medial aspect of right orbit right infratemporal fossa, right side anterior nasopharynx and soft palate, measuring approximately 9.1 X 6.7 X 8.2 cm (AP X TR X CC) with SUV max of 12. He was started on R-CHOP protocol. On the last follow up he is post 3 cycles of R-CHOP and the facial swelling has completely resolved, and he is clinically well.

Patient 3

46 year old, male presented with complaints of swelling of the right eye for 11 months, diplopia for 6 months, epiphora and mild pain in the right eye for 6 months. Rest system review was negative. On initial evaluation, imaging CECT orbit showed mildly enhancing soft tissue density in the right infraorbital region in the extraconal space. Biopsy of the right orbital mass was done and histopathological examination revealed low grade non-hodgkin lymphoma - extranodal marginal zone lymphoma. He was referred to SCI for further management. The positive examination findings included proptosis, epiphora and conjunctival congestion of the right eye. Rest examination was within normal limits. FDG PET CT whole body was done for staging and showed hypermetabolic soft tissue density measuring 2.9 x 4.7 x 3.9cm mass with SUV max of 8.42, encasing the extraocular muscles and partial encasement of the right optic nerve. PET CT also showed 1.1 x 0.7cm right parotid lymph node with mild metabolism. He was started on BR protocol. Post 2 cycles of chemotherapy the orbital swelling had completely resolved clinically. On last follow up he has completed 3 cycles of BR and is clinically well.

Patient 4

61 year old male, presented with complaints of swelling, redness, and lacrimation from left eye. Complaints had started 3 years back with swelling which gradually increased. He subsequently started to have redness and lacrimation. Rest system review was negative. He was evaluated for these complaints in February 2022. CE MRI orbit showed heterogeneously enhancing soft tissue component involving the medial wall and floor of left orbit with pre and post septal involvement and involvement of intra and extraocular muscles and optic nerve encasements with extensions into left premaxillary, pre zygomatic region, left buccal space, left masticator space and left parotid space and left submandibular region. He was started on steroids. Patient was lost to follow up till October 2024 till when he had continued steroids. He presented in October 2024 for progressive swelling and redness and watering form left eye. Repeat imaging CT Orbit shown enhancing infiltrative soft tissue density mass lesion in the inferior extraconal left orbit abutting the left lateral rectus, left inferior rectus, left inferior oblique muscles and left eye globe, causing proptosis of the left eye. The lesion erodes the bony floor of the left orbit with extension into the left infratemporal fossa. Left middle

cranial fossa meningeal thickening and enhancement is seen with reactive edema in the left temporal lobe,extending into the left masseteric space. Left pterygomaxillaryforamen, left spheno- orbital foramen and left foramen ovale, which is widened. Biopsy from the orbital mass on histopathological examination and immuno-histochemistry was suggestive of low grade B cell non Hodgkin lymphoma. Further tumour characterization was not done in view of exhaustion of tissue block and patient was not willing for a 2nd biopsy. He was started on BR protocol. In the last follow up he has completed 2 cycles of BR and is clinically improved.

DISCUSSION

In a major review of orbital lymphomas data of a total of 2,211 reported cases of orbital lymphoma were analyzed("Orbital Lymphoma," 2019) . The majority of cases were of B-cell origin, with 2,139 cases (97%). Seventy-two cases are of T-cell origin (3%), of which 46 cases are of mixed T/NK-cell origin (64%) ("Orbital Lymphoma," 2019). The most common B-cell lymphoma is extranodal marginal zone B-cell lymphoma, with 1,112 cases (59%) followed by diffuse large B-cell lymphoma , with 495 cases (23%) follicular lymphoma , with 182 cases (9%) and mantle cell lymphoma , with 99 cases (5%) ("Orbital Lymphoma," 2019) .The most common type of T-cell lymphoma is natural killer T-cell lymphoma, with 46 cases (64%) ("Orbital Lymphoma," 2019). Peripheral T-cell lymphoma not otherwise specified is the second most common T-cell lymphoma, with 8 cases (11%). In this case series we had 4 patients with orbital lymphoma with all 4 being B cell lymphomas. 2 patients were diagnosed with DLBCL, 1 patient was diagnosed with marginal zone lymphoma, and 1 patient was diagnosed with low grade B non-hodgkin lymphoma which was not further sub categorised.

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